
Reliable Classification of Functional Profiles and Movement Disorders of Children with Cerebral Palsy.

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Objective: To examine the inter-rater reliability of the Communication Function Classification System (CFCS), Bimanual Fine Motor Function (BFMF), Surveillance of Cerebral Palsy in Europe (SCPE) classification tree, and Gross Motor Function Classification System (GMFCS) in children with cerebral palsy (CP) and periventricular white matter injury (PWMI) aged 4-11 years. Method: Twenty children were assessed by two raters using the four tools, in addition parents undertook ratings on the Manual Ability Classification System (MACS). Kappa statistics were used to calculate the level of agreement between raters' classifications. Results: Participants comprised 12 males and 8 females with CP and PWMI, mean age 8 years 1 month (standard deviation 2 years 3 months). Interrater reliability across the four tools was 0.98 (CFCS, BFMF, and GMFCS) and 0.84 (SCPE). Implications: These findings suggest that these four tools are reasonably robust to inter-rater variability supporting their routine use along with the MACS in clinical and research applications.

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Reliability of "Modified timed up and go" test in children with cerebral palsy.

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BACKGROUND: Timed up and go (TUG) is a quick test used in clinical practice as an outcome measure to assess functional ambulatory mobility or dynamic balance in adults. However, little information is available on TUG test used in cerebral palsy. Hence, the purpose of our study was to assess the intra-rater reliability of TUG test in cerebral palsy children. AIM AND OBJECTIVE: To assess within-session and test-retest reliability after 1 week of TUG test in cerebral palsy children. Setting and Design: It was an a cross-sectional observational study conducted...
in a neurorehabilitation unit, with 30 cerebral palsy children of 4-12 years, within Gross Motor Function Classification System (GMFCS) level I, II, III, and with an IQ = 50. The sampling technique used was purposive sampling excluding children with cognitive deficit. MATERIALS AND METHODS: Subjects performed TUG on three occasions - Initial assessment (time 1), 30 min after initial assessment (time 2), and 1 week after initial assessment (time 3). Three trails were conducted for each of the three occasions. The mean score of three trials was documented as the final score. Within-session and test-retest reliability were analyzed using scores of time 1 and 2, and time 1 and 3, respectively. STATISTICAL ANALYSIS: The documented data were analyzed for within-session and test-retest reliability after 1 week of TUG test by using intraclass correlation coefficient (ICC). RESULTS: Reliability of TUG test was high, with ICC of 0.99 for within-session reliability and 0.99 for test-retest reliability. CONCLUSION: Intra-rater reliability of TUG test in cerebral palsy children was found to be high.

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AIM: The aim of this study was to examine the test-retest reproducibility (reliability and agreement) of the 6-minute push test (6MPT) and the one-stroke push test (1SPT), and construct validity of the 6MPT in children with cerebral palsy (CP) who self-propel a manual wheelchair. METHOD: Seventy-three children and adolescents with spastic CP (51 males, 22 females; mean age 11y 9mo, SD 3y 7mo, range 4-18y; three unilateral, 70 bilateral) using a manual wheelchair for at least part of the day were recruited from and tested in different rehabilitation settings in the Netherlands and Brazil. Participants were classified as Gross Motor Function Classification System (GMFCS) level II (n=7), III (n=36), or IV (n=30). RESULTS: Intraclass correlation coefficients (ICCs) for distance covered on the 6MPT (mean distance 266.5m, SD 120.6m) and the 1SPT (mean distance 4.5m, SD 2.7m) showed excellent reliability (ICC=0.97) for both tests. There was a significant correlation between the 6MPT and the 1SPT (r=0.73; p<0.001), and between the 6MPT and heart rate during the 6MPT (r=0.29, p=0.014). INTERPRETATION: These results indicate that both the 6MPT and the 1SPT test are reproducible functional tests for young people with CP who self-propel a wheelchair. Agreement for the 6MPT seems relatively large for children who perform short distances. Construct validity is supported for the 6MPT in children with CP.


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Are clinical measurements linked to the Gait Deviation Index in cerebral palsy patients?


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OBJECTIVE: From a dataset of clinical assessments and gait analysis, this study was designed to determine which of the assessments or their combinations would most influence a low gait index (i.e., severe gait deviations) for individuals with cerebral palsy. DESIGN: A retrospective search, including clinical and gait assessments, was conducted from August 2005 to September 2009. POPULATION: One hundred and fifty-five individuals with a clinical diagnosis of cerebral palsy (CP) (mean age (SD): 11 (5.3) years) were selected for the study. METHOD: Quinlan's Interactive Dichotomizer 3 algorithm for decision-tree induction, adapted to fuzzy data coding, was
employed to predict a Gait Deviation Index (GDI) from a dataset of clinical assessments (i.e., range of motion, muscle strength, and level of spasticity). RESULTS: Seven rules that could explain severe gait deviation (a fuzzy GDI low class) were induced. Overall, the fuzzy decision-tree method was highly accurate and permitted us to correctly classify GDI classes 9 out of 10 times using our clinical assessments. CONCLUSION: There is an important relationship between clinical parameters and gait analysis. We have identified the main clinical parameters and combinations of these parameters that lead to severe gait deviations. The strength of the hip extensor, the level of spasticity and the strength of the tibialis posterior were the most important clinical parameters for predicting a severe gait deviation.

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Challenges in organising surgical trials in cerebral palsy.

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PMID: 23266042 [PubMed - as supplied by publisher]


Rootless century: posterior rhizotomy for spastic cerebral palsy.

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Sudden onset odontoid fracture caused by cervical instability in hypotonic cerebral palsy.

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Fractures of the upper cervical spine rarely occur but carry a high rate of mortality and neurological disabilities in children. Although odontoid fractures are commonly caused by high-impact injuries, cerebral palsy children with cervical instability have a risk of developing spinal fractures even from mild trauma. We herein present the first case of an odontoid fracture in a 4-year-old boy with cerebral palsy. He exhibited prominent cervical instability due to hypotonic cerebral palsy from infancy. He suddenly developed acute respiratory failure, which subsequently required mechanical ventilation. Neuroimaging clearly revealed a type-III odontoid fracture accompanied by anterior displacement with compression of the cervical spinal cord. Bone mineral density was prominently decreased probably due to his long-term bedridden status and poor nutritional condition. We subsequently performed posterior internal fixation surgically using an onlay bone graft, resulting in a dramatic improvement in his respiratory failure. To our knowledge, this is the first report of an odontoid fracture caused by cervical instability in hypotonic cerebral palsy. Since cervical instability and decreased bone mineral density are frequently associated with cerebral palsy, odontoid fractures should be cautiously examined in cases of sudden onset respiratory failure and aggravated weakness, especially in hypotonic cerebral palsy patients.

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The aim of this study was to develop and evaluate an instrument - the Responsive Augmentative and Alternative Communication Style (RAACS) scale Version 2 - to assess the communicative style of parents as they interact with their children using augmentative and alternative communication (AAC). This scale was used to analyze play interactions between 43 parents and 28 children with different diagnoses (including Down syndrome, autism, cerebral palsy, and intellectual disability), aged between 12 and 60 months. Parent-child interactions were observed both before and after parent participation in ComAlong, a training course on using responsive communication and AAC to support interaction with children. Based on an analysis of the results, Version 3 of the RAACS scale was developed and is recommended for future use. Analyses of Version 3 showed acceptable inter- and intra-coder reliability, and excellent internal consistency.

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The association of drooling and health-related quality of life in children with cerebral palsy.

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OBJECTIVE: To investigate the association between drooling in children with cerebral palsy (CP) and their health-related quality of life (HRQOL), as well as the possible variables that predict their HRQOL. METHOD: A cross-sectional design was used for this study. Children with CP, without other identified disease, aged 2 to 6 years, who drool (n = 33) or did not drool (n = 14), were included. The dependent variables were the physical health summary scores and the psychosocial health summary scores of the Pediatric Quality of Life Inventory version 4.0. The t test, Pearson product-moment correlation, Mann-Whitney U test and stepwise regression analysis were used for statistical analysis. RESULTS: The physical health and psychosocial health summary scores of the children that drooled (16.29 ± 15.97 and 42.92 ± 17.57, respectively) were lower than for the children that did not drool (31.97 ± 22.22 and 57.09 ± 12.21, respectively; P < 0.01). The drooling ranking score was negatively correlated with the physical health summary score (r = -0.355; P < 0.05) and the psychosocial health summary score (r = -0.381; P < 0.01). The stepwise regression showed that gross motor development and the drooling ranking score predicted 56.6% of the variability of the physical health summary score (R(2) = 0.566; P < 0.01). The language development score predicted 25.6% of the variability of the psychosocial health summary score (R(2) = 0.256; P < 0.01). CONCLUSION: Drooling was associated with a lower HRQOL. Prediction of the physical health summary score was more closely associated with gross motor development and the drooling ranking scores. Prediction of the psychosocial health summary score was more closely associated with the language development of children with CP aged 2 to 6 years.

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Digestive tract neural control and gastrointestinal disorders in cerebral palsy.

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OBJECTIVES: To examine the neural control of digestive tract and describe the main gastrointestinal disorders in cerebral palsy (CP), with attention to the importance of early diagnosis to an efficient interdisciplinary treatment.

SOURCES: Systematic review of literature from 1997 to 2012 from Medline, Lilacs, Scielo, and Cochrane Library databases. The study included 70 papers, such as relevant reviews, observational studies, controlled trials, and prevalence studies. Qualitative studies were excluded. The keywords used were: cerebral palsy, dysphagia, gastroesophageal reflux disease, constipation, recurrent respiratory infections, and gastrostomy.

SUMMARY OF THE FINDINGS: The appropriate control of the digestive system depends on the healthy functioning and integrity of the neural system. Since CP patients have structural abnormalities of the central and peripheral nervous system, they are more likely to develop eating disorders. These range from neurological immaturity to interference in the mood and capacity of caregivers. The disease has, therefore, a multifactorial etiology. The most prevalent digestive tract disorders are dysphagia, gastroesophageal reflux disease, and constipation, with consequent recurrent respiratory infections and deleterious impact on nutritional status.

CONCLUSIONS: Patients with CP can have neurological abnormalities of digestive system control; therefore, digestive problems are common. The issues raised in the present study are essential for professionals within the interdisciplinary teams that treat patients with CP, concerning the importance of comprehensive anamnesis and clinical examination, such as detailed investigation of gastrointestinal disorders. Early detection of these digestive problems may lead to more efficient rehabilitation measures in order to improve patients' quality of life.

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Acute functional deterioration in a child with cerebral palsy.

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We describe a case of acute functional deterioration in a 13-year-old girl with severe spastic diplegia (GMFCS III) and a new diagnosis of diffuse intrinsic pontine glioma (DIPG). She presented with acute deterioration in mobility and motor function over 1 month, which was associated with dysarthria, dysphagia and behavioural change. Her mother had noticed subtle functional deterioration over the 2 months prior to this. Her physiotherapist who was concerned about her acute functional deterioration referred her for emergency review. Neurological imaging revealed a diffuse pontine lesion consistent with DIPG. She was subsequently referred to oncology. She deteriorated further, clinically, over the next few days and following discussion with the team; her family opted for palliative treatment, given the poor prognosis associated with DIPG.

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Inpatient Growth and Resource Use in 28 Children's Hospitals: A Longitudinal, Multi-institutional Study.

Berry JG, Hall M, Hall DE, Kuo DZ, Cohen E, Agrawal R, Mandl KD, Clifton H, Neff J.

OBJECTIVE To compare inpatient resource use trends for healthy children and children with chronic health conditions of varying degrees of medical complexity. DESIGN Retrospective cohort analysis. SETTING Twenty-eight US children's hospitals. PATIENTS A total of 1 526 051 unique patients hospitalized from January 1, 2004, through December 31, 2009, who were assigned to 1 of 5 chronic condition groups using 3M's Clinical Risk Group software. INTERVENTION None. MAIN OUTCOME MEASURES Trends in the number of patients, hospitalizations,
hospital days, and charges analyzed with linear regression. RESULTS Between 2004 and 2009, hospitals experienced a greater increase in the number of children hospitalized with vs without a chronic condition (19.2% vs 13.7% cumulative increase, P < .001). The greatest cumulative increase (32.5%) was attributable to children with a significant chronic condition affecting 2 or more body systems, who accounted for 19.2% (n = 63 203) of patients, 27.2% (n = 111 685) of hospital discharges, 48.9% (n = 1.1 million) of hospital days, and 53.2% ($9.2 billion) of hospital charges in 2009. These children had a higher percentage of Medicaid use (56.5% vs 49.7%; P < .001) compared with children without a chronic condition. Cerebral palsy (9179 [14.6%]) and asthma (13 708 [21.8%]) were the most common primary diagnosis and comorbidity, respectively, observed among these patients.

CONCLUSIONS Patients with a chronic condition increasingly used more resources in a group of children's hospitals than patients without a chronic condition. The greatest growth was observed in hospitalized children with chronic conditions affecting 2 or more body systems. Children's hospitals must ensure that their inpatient care systems and payment structures are equipped to meet the protean needs of this important population of children.

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The changes in the brain's electrical activity in children with cerebral palsy during the complex rehabilitation [Article in Russian]

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One hundred and five children, aged from 3 to 7 years, with the diagnosis "spastic diplegia cerebral palsy" were treated. Patients were stratified into three groups: group I (n=36) received three courses of microcurrent therapy (MENS) in addition to standard treatment; group II (n=38) received three courses of MENS in the combination with two treatment courses with the nootropic drug cortexin; children of group III (n=31) received standard therapy using massage and gymnastics. MENS was carried out in courses, including 15 sessions each, using the apparatus "MEKS". Cortexin was introduced intramuscular in dosage 10 mg, the treatment course consisted of 10 injections. To the end of the rehabilitation program, positive changes were found: 50% patients of group I, 66% patients of group II and 16% patients of group III could perform complex instructions and acquired skills in modeling and recognition of geometric forms. Positive changes in the brain's electrical activity were found in 75% of children in group I, in 82% of children in group II and in 64% of children in group III.

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Therapeutic potential of autologous stem cell transplantation for cerebral palsy.

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Background. Cerebral palsy (CP) is a severe disabling disease with worldwide incidence being 2 to 3 per 1000 live births. CP was considered as a noncurable, nonreparative disorder, but stem cell therapy offers a potential treatment for CP. Objective. The present study evaluates the safety and efficacy of autologous bone-marrow-derived mononuclear cell (BMMNCs) transplantation in CP patient. Material and Methods. In the present study, five infusions of autologous stem cells were injected intrathecally. Changes in neurological deficits and improvements in function were assessed using Gross Motor Function Classification System (GMFCS-E&R) scale. Results. Significant motor, sensory, cognitive, and speech improvements were observed. Bowel and bladder control has been achieved. On the GMFCS-E&R level, the patient was promoted from grade III to I. Conclusion. In this study, we report that intrathecal infusion of autologous BMMNCs seems to be feasible, effective, and safe with encouraging functional outcome improvements in CP patient.

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DTI correlates of cognition in term children with spastic diplegic cerebral palsy.


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BACKGROUND AND AIMS: Presently, there is no published study that shows association between cognition and white matter injury in spastic cerebral palsy. We aimed to correlate cognitive functions with diffusion tensor imaging derived metrics in spastic diplegic children. METHODS: Twenty-two term children with spastic diplegia and 22 healthy controls were recruited. All patients were graded on the basis of gross motor function. The Indian children intelligence Test was used to quantify cognition and diffusion tensor imaging was used to quantify microstructural changes in various white matter regions. Diffusion tensor imaging metrics were quantified by placing regions of interests in different white matter regions like corona radiata, anterior limb of internal capsule, posterior limb of internal capsule, mid brain, pons, medulla, genu, splenium, temporal white matter, parietal white matter, frontal white matter and occipital white matter. RESULTS: Spastic diplegic children showed significantly lower neuropsychological test scores as compared to controls. A significantly decreased fractional anisotropy values were observed in corona radiata, anterior limb of internal capsule, posterior limb of internal capsule, mid brain, pons, medulla, genu, splenium and occipital white matter; however significantly increased mean diffusivity values were observed in corona radiata, anterior limb of internal capsule, posterior limb of internal capsule, mid brain, pons and genu in spastic diplegic as compared to controls. A significant positive correlation in fractional anisotropy and negative correlation in mean diffusivity was observed with neuropsychological test scores. CONCLUSION: These results suggest that these imaging metrics may be used as a biomarker of cognitive functions in term children with spastic diplegia.

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Population study of neurodevelopmental outcomes of extremely premature infants admitted after office hours.

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AIM: The aim of the study was to compare neurodevelopmental outcomes of extremely preterm infants admitted during (OH) and after (AH) office hours. METHODS: A retrospective review of the New South Wales and Australian Capital Territory Neonatal Intensive Care Units’ (NICUs) Data Collection of all infants <29 weeks gestation admitted to New South Wales and Australian Capital Territory NICUs between January 1998 and December 2004 was conducted. The primary outcome was moderate/severe functional disability (FD) at 2-3 years follow-up defined as developmental delay (Griffiths Mental Developmental Scales general quotient or Bayley Scales of Infant Development-II mental developmental index >2 standard deviations below the mean), cerebral palsy (unable to walk without aids), deafness (requiring bilateral hearing aids) or blindness (visual acuity <6/60 in the better eye). RESULTS: Mortality and age at follow-up were comparable between the AH and OH groups. Developmental outcome was evaluated in 972 (74.9%) infants admitted during AH and 501 (74.6%) admitted during OH. FD was not significantly different between the AH and OH groups (17.1% vs. 14.8%, adjusted odds ratio 1.131, 95% confidence interval 1.131 (0.839-1.523), P = 0.420). There were no significant differences between AH and OH infants with cerebral palsy (9.6% vs. 7.6%), developmental delay (5.4% vs. 5.0%) or any other component of FD. CONCLUSION: There is little circadian variation in mortality and adverse neurodevelopmental outcomes in an NICU network with the current model of after hours staffing and support, and sharing of NICU workload within a network.


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Obstetric outcomes in women who sustained a spinal cord injury during pregnancy.

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Study design: Case report. Setting: Prince of Wales Spinal Cord Injuries Unit, Sydney, Australia. Methods: Interrogation of our unit database identified only two women who became spinal cord injured while pregnant; their medical records were reviewed and an unstructured follow-up telephone interview conducted 6 years after discharge. Case 1: CC sustained a fracture dislocation with paraplegia at the sixth thoracic level (T6) in a motorbike accident while she was pregnant, 12-week gestational age (GA). Profound shock and hypoxia complicated the injury and recurrent urinary tract infections complicated the rest of her pregnancy. A baby with arthrogryposis multiplex congenita was delivered at full term. Severe cerebral palsy (CP) and deafness were present at follow-up 6 years later. Case 2: A 33-year-old multigravida, 27 weeks GA, developed sudden, spontaneous onset of paraplegia (T3 ASIA B) due to an extradural haematoma, which was evacuated on the day of admission. Systolic blood pressure was maintained above 90?mm?Hg during and after surgery. A normal, healthy boy was delivered by caesarean section at 40 weeks GA and remained so at 6 years. Conclusion: Traumatic spinal cord injury (SCI) with its attendant multiple potential insults to the developing foetus results in a high risk of foetal loss and malformation particularly in the first trimester. However, if the injury occurs later in pregnancy and if blood pressure and oxygenation are maintained, the risk of foetal loss and abnormality may be substantially reduced. Spinal Cord advance online publication, 18 December 2012; doi:10.1038/sc.2012.125.

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Phenomenon of cell genome instability in the pathogenesis of children cerebral palsy [Article in Russian]
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Perinatal and neonatal predictors of developmental coordination disorder in very low birthweight children.
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OBJECTIVE: To identify perinatal and neonatal risk factors associated with developmental coordination disorder (DCD) in very low birthweight children (VLBW: <1250 g). DESIGN: Retrospective design with prospectively collected cohort. SETTING: Neonatal Follow-Up Program, Vancouver, Canada. PATIENTS: 157 VLBW children assessed at 4-5 years who were free of cerebral palsy or major neurological impairment and had full-scale IQ >70. MAIN OUTCOME MEASURE: Movement Assessment Battery for Children (MABC). ESULTS: Using ≥15th percentile on the MABC as the cut-off, 42% of our cohort developed DCD. Perinatal variables significantly associated with DCD were male sex, lower gestational age and lower birth weight, but only male sex and low birth weight independently predicted DCD, accounting for 20% of the variance in MABC scores. Compared with children without motor impairment, children with DCD had greater postnatal steroid exposure, longer duration of ventilation, more days on oxygen and significant retinopathy of prematurity, but only postnatal steroid exposure was significant, accounting for an additional 3% of the variance in MABC scores. Boys performed more poorly than girls on all subtests of the MABC. CONCLUSIONS: Male sex and low birth weight were significant predictors of DCD, suggesting that these infants should be followed for detection of this common, but under-recognised disorder. Future research aimed at identifying neural underpinnings of DCD and possible antecedents to the disorder is warranted.

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Gene expression in archived newborn blood spots distinguishes infants who will later develop cerebral palsy from matched controls.
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Background: Gene expression in archived newborn blood spots remaining from newborn screening may reflect pathophysiological disturbances useful in understanding the etiology of cerebral palsy (CP). Methods: We quantified the expression of gene sets representing four physiological pathways hypothesized to contribute to CP in archived unfrozen residual newborn blood spot specimens from 53 children with CP and 53 age, gender, and gestational-age matched controls. We selected four empirical and three canonical gene sets representing inflammatory, hypoxic, coagulative, and thyroidal pathways, and examined mRNA expression using an 8x60K oligonucleotide microarray. The log(2) fold change of gene expression between matched cases and controls were analyzed using the Generally Applicable Gene Set Enrichment (GAGE) method. Results: The empirical inflammatory and empirical hypoxic gene sets were significantly down-regulated in term-born CP cases (N = 33) as compared to matched controls (P = 0.0007 and 0.0009, respectively), while both gene sets were significantly up-regulated (P = 0.0055 and 0.0223, respectively) in preterm-born CP cases (N = 20). The empirical thyroidal gene set was significantly up-regulated in preterm-born CP (P = 0.0023). Conclusion: The newborn blood spot
transcriptome can serve as a platform for investigating distinctive gene expression patterns in children who later develop CP.

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